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CLINICAL IMAGE

Acute liver failure revealing hepatic vasculitis associated to SLE

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Presentation

A 43-year-old man on anticoagulation for 23 years for a triple-positive venous thrombotic antiphospholipid syndrome (APS) considered as isolated, finally presented with a first systemic lupus erythematosus (SLE) flare, associating arthralgias, class III + V lupus nephritis, Evans syndrome and anti-dsDNA antibodies. On day third of intravenous methylprednisolone, he declared fever, upper right quadrant pain and acute liver failure. Doppler ultrasonography was unremarkable, but hepatobiliary MRI showed multiple nodular lesions (arrows): enhanced in T1 sat fat (Fig. 1a) after contrast injection, DWI hypersignal (Fig. 1b) and T2 hypersignal (Fig. 1c). Targeted liver biopsies showed a diffuse neutrophilic inflammatory infiltrate with some Councilman bodies, parietal necrosis of centro-lobular veins (Fig. 1d, arrow) and an enlargement of portal spaces with fibrinoid necrosis of the portal venule (Fig. 1e, arrow). ANCA and cryoglobulinemia were negative; infections were ruled out. A diagnosis of hepatic SLE-associated vasculitis (SAV) was retained. Rituximab added to corticosteroids and hydroxychloroquine allowed a rapid improvement of liver function and Evans syndrome along with MRI normalization three months later. MMF was started as maintenance treatment without SLE relapse after 3 years of follow-up.

Discussion

SAV occurs in 11% to 36% SLE patients and mainly involves small vessels, especially post-capillary venules (80–90%) [1, 2]. SAV mostly affect the skin. Few patients exhibit an internal organ vasculitis, and of these, hepatic involvement has rarely been reported. In this settings, biopsy examination of the involved tissue is essential to confirm vasculitis and also look for associated diagnoses, such as APS-related microangiopathy [3]. Finally, in this case rituximab has been effective to treat hepatic SAV.

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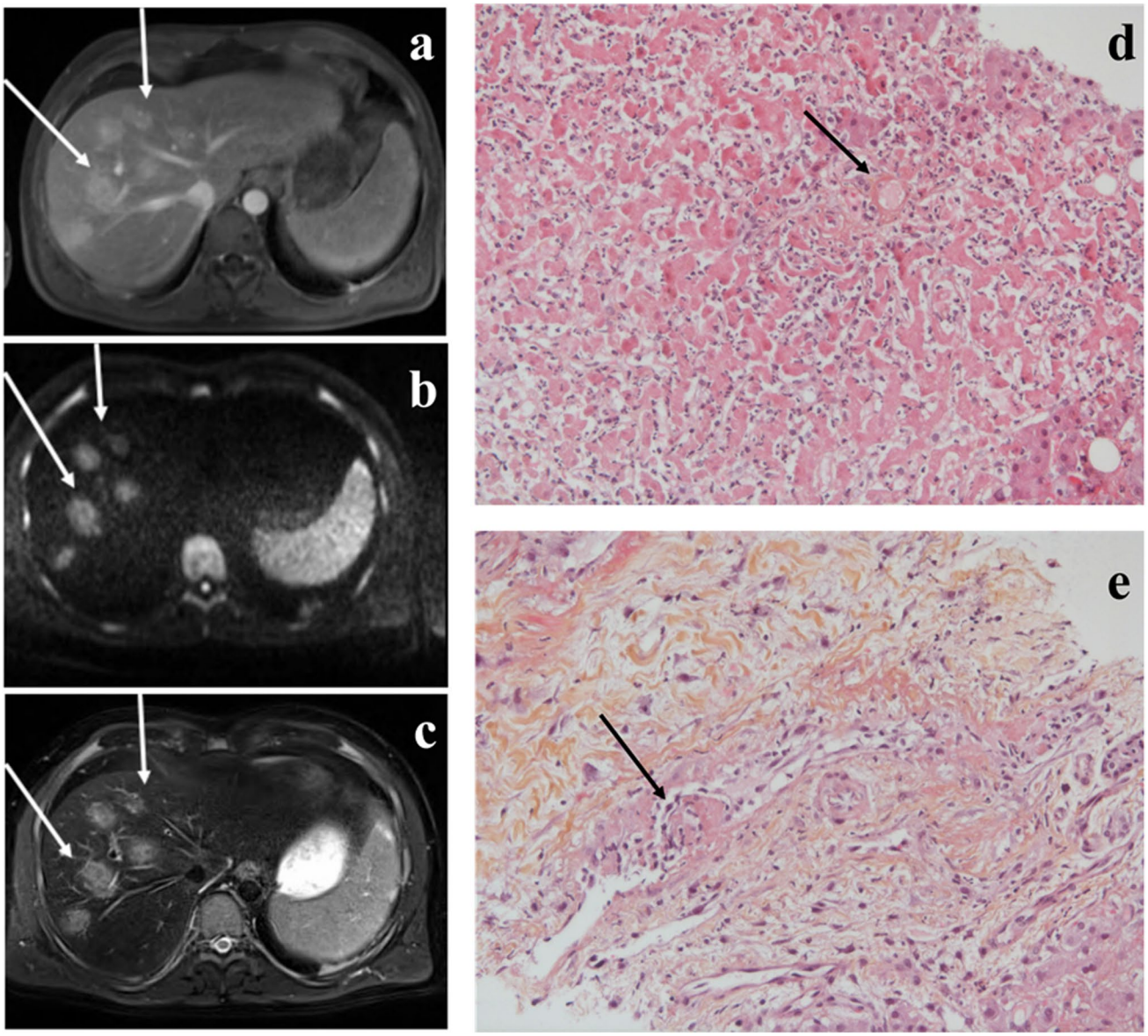

Fig. 1 Hepatobiliary MRI showing multiple nodular lesions (arrows). **a** Axial T1 sat fat sequence showing enhancement of nodular lesions after contrast injection. **b** Axial DWI sequence showing hypersignal of nodular lesions. **c** Axial T2 showing hypersignal of nodular lesions. Targeted liver biopsy. **d** Infiltration of the hepatic lobular parenchyma by a diffuse inflammatory infiltrate of neutrophils. Presence of some Councilman bodies. Parietal necrosis of a centro-lobular vein (arrow) (Hemalun eosin saffron stain, magnification × 20). **e** Enlargement of a portal space with fibrinoid necrosis of the portal venule (arrow) (Hemalun eosin saffron stain × 40)

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Declarations

Ethics approval N/A.

Consent for publication We obtained written informed consent from the patient for publication of this case report together and any accompanying images.

Disclosures None.

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